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# SOLITARY ANGIOKERATOMA OF THE TONGUE MASQUERADING AS PAPILLOMA – A RARE CASE REPORT AND LITERATURE REVIEW

**Oral Pathology**


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## ABSTRACT

The presence of dilated and congested blood vessels in the superficial dermis, accompanied by thickening of the stratum corneum and the prickly layer characterizes angiokeratomas. Solitary angiokeratomas of the tongue are an exceptionally rare phenomenon, with fewer than 10 documented cases in the English medical literature. This report describes a case of this unusual presentation in a 10-year-old boy, initially misdiagnosed as papilloma on clinical examination. Incisional biopsy initially suggested lymphangiomatous hamartoma, while the definitive diagnosis of solitary angiokeratoma of the tongue was established only with excisional biopsy.



## KEYWORDS

Angiokeratoma, Tongue papilloma, Vascular malformation, Oral lesion


**INTRODUCTION:**

Tongue angiokeratomas represent a highly unusual presentation, with documented instances being exceptionally rare.[1] On histopathological examination of tissue samples, angiokeratomas reveal widened blood channels within the underlying skin (dermal ectasia) associated with increased thickness of the outermost skin layers (hyperkeratosis and acanthosis) and deeper indentations of the skin surface (elongated rete ridges).[1-7] Angiokeratomas encompass five distinct subtypes, varying in their clinical presentation while sharing a consistent pattern under microscopic examination.

**Case Report**

A 10-year-old boy presented with a four-month history of a gradually enlarging swelling on the right posterior third of his tongue. Initially noted as a small lesion, it progressively increased in size and expanded towards the tongue base. Trauma from a tongue bite was reported, but no fever, slurred speech, difficulty swallowing, or spontaneous bleeding from the lesion were present. The patient denied any similar lesions elsewhere and had no notable medical or family history. No prior treatment for the swelling had been sought.

Oral examination revealed a papulonodular mass measuring approximately 4x2 cm on the right posterior third of the tongue, extending towards the base. The lesion displayed a blueish-brown hue, soft consistency, granular texture, and lacked tenderness or bleeding with gentle palpation. No abnormalities were observed in the remaining oral mucosa. Vital signs remained stable, and a comprehensive systemic examination yielded no further cutaneous or systemic anomalies. Based on clinical presentation, the provisional diagnosis was papilloma of the tongue.

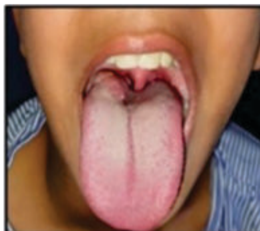

**Figure1:** A papulonodular growth on right posterior one-third of tongue extending towards the base of the tongue.

Routine hematologic and biochemical tests yielded normal results. An initial incisional biopsy was performed, revealing a thinned squamous epithelium overlying dilated, thin-walled vascular channels in the subepithelial tissue. These channels, lined by endothelial cells and filled with lymph, compressed and extended into the epidermis, suggesting the possibility of lymphangioma.

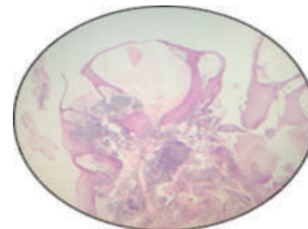

**Figure 2:** Histopathological examination of Incisional biopsy from the lesion showing dilated thin-walled channels filled with lymph compressing and extending into epidermis.

Following surgical excision, the lesion was preserved in formalin and sent for microscopic examination. The excised tissue appeared greyish-brown and soft in consistency and measured 3.8 x 1.5 x 0.6 cm. Microscopic examination revealed prominent dilation of blood vessels within the papillary dermis, extending partially into the epidermis. The overlying epidermis exhibited papillomatous hyperplasia, thinned out above ectatic vascular structures, alternating with thickened layers (acanthosis with orthokeratosis) and deeper invaginations (elongation of rete ridges). Notably, no atypical or malignant features were identified in the examined sections. Based on these findings, the definitive diagnosis was established as inflamed angiokeratoma of the tongue.

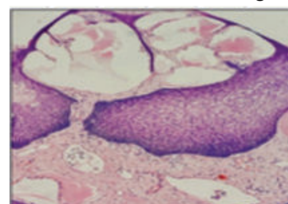

**Figure 3**

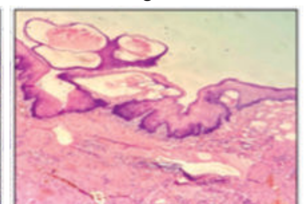

**Figure 4**

**Figure 3&4:** Vascular ectasia seen in papillary dermis also extending

into surface epithelium. Overlying stratified squamous epithelium showed hyperplasia, orthokeratosis, and elongation of rete ridges.

**Discussion and Review of Literature**

Often symptomless in their early stages, angiokeratomas may manifest with mild discomfort as they enlarge. Despite their rarity and typically congenital nature, some cases may develop in early childhood or even adulthood.[4-10] While angiokeratomas can develop in otherwise healthy individuals with no underlying conditions, they can also be associated with specific inherited enzyme deficiencies like *Anderson-Fabry disease* (a rare X-linked lysosomal storage disorder).[3-7]

Similarly, they can coexist with *Nevus flammeus* (birthmarks) and *Cavernous hemangiomas* (vascular malformations).[2] Interestingly, while cutaneous angiokeratomas are more commonly observed in males, tongue angiokeratomas exhibit a female predilection, with a three-to-one female-to-male ratio.[1,3] However in our case, it was a young male patient.

Our understanding of angiokeratoma's origin remains somewhat shrouded in mystery. One prevailing theory suggests it emerges as a telangiectatic lesion, stemming from capillary damage in the papillary dermis. This damage, triggered by local trauma or increased venous pressure, might subsequently induce secondary epithelial proliferation. Alternatively, some experts propose that angiokeratomas arise due to enhanced proliferative potential alongside the pre-existing vascular malformation and the close proximity of epidermis to vascular spaces, favouring reactive epidermal growth.[3]

Angiokeratoma can also develop in an area of lymphangioma circumscriptum after repeated local injuries. Despite initial suspicion of lymphangioma based on the incisional biopsy from the tongue, the case took an unexpected turn. Histopathological examination of the subsequent excisional biopsy revealed features consistent with inflamed angiokeratoma, highlighting the potential for diagnostic challenges and the importance of thorough evaluation.

Clinically, five distinct types of angiokeratoma can be identified: *Angiokeratoma of Fordyce*, primarily confined to the scrotum and vulva; *Angiokeratoma of Mibelli*, predominantly found on the backs of fingers and toes; *Angiokeratoma corporis diffusum,* characterized by a bathing suit distribution and often linked to Anderson-Fabry disease; *Angiokeratoma circumscriptum*, the least common type, which may co-occur with nevus flammeus, cavernous hemangioma, or present as solitary angiokeratoma or multiple lesions of unknown cause.[3,7]

Although they have different clinical presentations but they are histologically identical. Based on the clinical details, this case is idiopathic solitary angiokeratoma of tongue.

Angiokeratoma circumscriptum of the tongue can share similarities with various other lesions, necessitating careful differentiation. Key contenders include lymphangioma circumscriptum, hemangioma, verruca vulgaris, melanocytic nevus, malignant melanoma, capillary aneurysm, cystic hygroma, and papilloma.[2,3,4] CD34 antigen positivity can be used to confirm the diagnosis [3].

Given the benign and typically asymptomatic nature of angiokeratoma, treatment isn't always an immediate priority. However, intervention may be considered if the patient experiences discomfort, bleeding, or cosmetic concerns. Surgical excision and laser ablation remain the standard approaches, both demonstrating high efficacy.[1,4]

**Table 1: Summary of Literature Review**

| Author | Age/Sex | Site of lesion | Associated disease | Type of Angiokeratoma | Treatment | Recurrence | Malignant transformation |
|---|---|---|---|---|---|---|---|
| Kar HK, Gupta L1 | 12yr/M | Ventral aspect of tongue | None | Angiokeratoma circumscriptum | Combination of $CO_2$ and PDL | None | Nil |
| Kumar KS et al 2 | 11Yr/M | Dorsum of tongue | None | Solitary Angiokeratoma | Surgical excision | None | Nil |
| Kandalgaonkar S et al 3 | 38Yr/M | Tip of the tongue | None | Solitary Angiokeratoma | Surgical excision | None | Nil |
| Adorisio O et al 4 | 13Yr/M | Posterior part of the tongue | Cystic Fibrosis | Solitary Angiokeratoma | Surgical excision | None | Nil |
| Dutta M et al 5 | 18Yr/M | Base of the tongue | None | Isolated Angiokeratoma | Surgical excision | None | Nil |
| Job AM et al 6 | 12Yr/M | Dorsum of the tongue | None | Solitary Angiokeratoma | Surgical Excision | None | Nil |
| Giordano L et al 7 | 67yr/M | Posterior margin of tongue | None | Solitary Angiokeratoma | Excision | None | Nil |

**CONCLUSION:**

Solitary oral angiokeratoma, though infrequent, poses a diagnostic challenge. A thorough investigation for underlying congenital or acquired systemic conditions and any similar lesions elsewhere is critical. Definitive diagnosis hinges on surgical excision and histopathological examination. Further research is warranted to shed light on the potential for malignant transformation and recurrence in such cases.

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None

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